A CASE OF PROGRESSIVE NEUROTIC MUSCULAR ATROPHY.

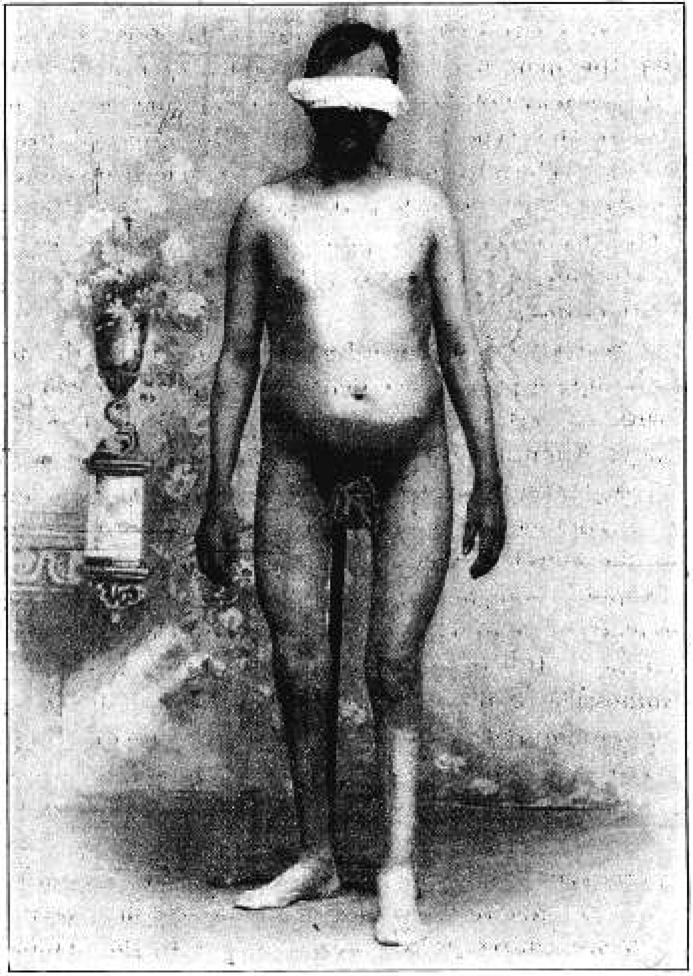
By CHARLES W. BURR, M.D.

History.—L. A., male, 31 years old, a clerk, applied for treatment at the Medico-Chirurgical Hospital in October, 1896, complaining of a slowly increasing difficulty in walking and weakness in the hands. His family history is negative. His previous health has always been good. He has not had gonorrhea or syphilis, and has never used alcohol. His sexual habits are good.

The present trouble began when he was in his twenty-sixth year, with severe pain in the left heel soon extending to the left calf. In three or four weeks his left ankle began to be weak, and pain came on in the left knee and hip. A year later he noticed weakness of the right lower limb without preceding pain. A few months after the onset he noted wasting in the left calf, and later (after the weakness) in the right. Wasting and weakness of the hands began several years after the legs were affected, and was not preceded or accompanied by pain or paresthesia. He has never had any loss of control of the bladder or rectum. His sexual power is good.

Examination.—The gait is waddling. He cannot stand with the feet close together, because of weakness. He cannot extend the toes, and can extend the feet only weakly. There is neither club foot nor other deformity. There is marked weakness in all the movements of the hands. Extension of the forearms is good, but flexion is rather weak. The shoulder and hip movements are normal. There is great wasting of both calves, some of the thighs, especially the left, and a little of the intrinsic muscles of the feet. The muscles of the hands are much wasted, the forearms somewhat, the upper arms little, if any; and the shoulders, neck and face not at all. The knee, biceps tendon, cremasteric, and muscle jerks are absent. There are no fibrillary twitchings. Sensation of touch, pain and temperature is normal. There are no vasomotor symptoms.

The first description of progressive, neurotic atrophy was given by Charcot and Marie in a paper in the Revue de Médecine for February, 1886. A short time later Howard Tooth published a thesis describing the same condition, entitled: "The Peroneal Type of Progressive Muscular Atrophy." He quoted cases which had been previously reported by Friedreich, Eichhorst and others, without their having recognised the nature of the affection. According to Charcot and Marie, the disease is characterized by muscular atrophy attacking first the feet and calves, and after considerable time, even several years, affecting the hands and the forearms, but never the trunk nor face. Fibrillary contractions may be present. There are sometimes vasomotor symptoms. Sensation is most often intact, but may be altered in several ways. Reaction of degeneration is frequent. The disease usually begins in early life, often affects several brothers and sisters, and may appear in succeeding generations. Clinical cases have been reported in Europe by Hoffmann, Bernhardt, Vizioli, Donkin, Dejerine and others. The descriptions agree in the main, though differing in detail. For example, Hoffmann has shown that the wasting may appear in the hands first, the legs being affected later. Sensory symptoms may be slight or severe. Thus, Sachs, of New York, says: "Sensory changes are generally present. The various forms of sensation may be slightly altered, or, in some cases, tactile sensation and temperature sense may remain normal, while the pain sense may be more distinctly involved. Paresthesia may be present in addition to the objective changes in sensation." Gowers states that cutaneous sensibility, though often normal, is sometimes impaired or lost, especially over the region in which the atrophy is greatest, or upon the soles of the feet. Pains occur in some cases.



The electrical reactions of the muscles vary, depending in large degree upon the stage during which the examination is made. There may be complete absence of electrical response, or partial or complete reaction of degeneration, or a mere quantitative diminution.

The only cases reported in America are two described by Sachs, one shown at the November meeting of this Society by Potts, and one not yet reported, to be presented here to-night by Dercum and Leopold, in all, counting this man, five. The Farr family, described by Osler, may, or may not, belong to this group. They were reported before this type of atrophy was distinguished from the others. The affection described by G. Dejerine and Sottas in 1893 (Mém. de la Soc. de Biologie), and G. Dejerine in 1896 (Revue de Méd.), under the title, "Interstitial, Hypertrophic and Progressive Neuritis of Childhood," is a distinct disease.

Scarcely the first word has been said about the morbid anatomy of progressive neurotic atrophy, nor can we say with any certainty that it is due to the same lesion in all cases. Clinically, it is an entity; pathologically, it probably is not. Muscular atrophy must, so far as present knowledge goes, be due to diseases of the spinal cord, the nerves, or the muscles, since we may disregard the rare cases of atrophy accompanying cortical disease, as being as yet unexplained. Often we can differentiate the three kinds of atrophy. But types shade into each other, and it is often impossible to tell whether a given case be due to nerve disease, muscular disease, or even cord disease. But few necropsies have been made in cases of progressive neurotic atrophy. In the early cases of Virchow and Friedreich, published before the clinical recognition of a distinct neural type of atrophy, there was found degeneration of the nerves and of the posterior columns of the spinal cord, especially affecting the columns of Goll. In 1890 Dubreuilh (Revue de Médecine, Vol. X.) reported a case of a child whose mother and eleven brothers and sisters suffered from the same disease. The patient became ill during the second year with a gradually increasing atrophic palsy, which began in the hands and later spread to the legs. There were no sensory disturbances. Death was caused by tuberculosis. Post-mortem there was found degeneration of the nerves, evidently of long standing, affecting especially the motor nerves of the hands and feet, and diminishing in intensity as the cord was approached. There was a very slight increase of the neuroglia in the columns of Goll and in the lower part of the columns of Burdach. The gray substance was normal. The changes in the muscles consisted of simple atrophy of the fibres, with, in some, a loss of the transverse striations and a proliferation of the nuclei. Some of the fibres were degenerated, a few hypertrophied. This, therefore, was a genuine case. But in 1896 Oppenheim and Cassirer (Deutsche Zeitschrift für Nervenheilkunde) reported a case which shows well the great liability to error in the pathological diagnosis of this affection. The patient was a man forty-two years old, and with good inheritance. Within two years an atrophic palsy, accompanied by pain, had developed. The distal parts of the extremities were affected---in the legs the muscles supplied by the peroneal nerves especially and in the arms the small muscles of the hands, and later the triceps and long supinator muscles. There was incomplete reaction of degeneration, and in some muscles no electrical reaction. Sensation was normal. The orbicularis palpebrarum was affected, but no other face muscle. The disease developed slowly, but progressively. Death resulted from angina. During life the case was diagnosticated as either a chronic multiple neuritis or neurotic muscular atrophy. The necropsy revealed only muscular disease. Microscopic examination of the intramuscular nerves, the nerve trunks, and the central nervous system showed practically no pathological condition. The muscles on the other hand showed the changes found in primary myopathies, namely, atrophy of the muscle fibres with occasional hypertrophy, absence of the transverse striation, with the appearance of longitudinal striation, partial vacuolation, great increase in the connective tissue and lipomatosis. These are not pathognomonic of myopathy, but are characteristic. Hoffmann asserts that there is a primary ascending degeneration of the nerve fibres, which secondarily involves the spinal cord, especially the columns of Goll and the motor cells of the anterior horns.

All things considered, we are probably justified in concluding that disease either of the muscles themselves, or of the nerves, may produce the clinical picture of progressive neurotic muscular atrophy, and that only in individual cases can we, by the accompanying symptoms, the electrical reaction, the presence or absence of sensory and vasomotor symptoms and the like make a correct pathologic diagnosis.

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Paralysis of the Ocular Muscles due to Lead Poisoning.

D. Casey Wood is reported (Journal of the American Medical Association; Columbus Medical Journal) to have presented the following case to the Chicago Ophthalmological Society (Feb. 9th, 1897):

Patient, aged twenty-nine, a painter, had lead colic five years ago, followed in two years by an attack of vertigo coming on suddenly one morning, and was then unconscious for half an hour, and had pain through his head for two days, at the end of which time he complained of diplopia and in three weeks noticed that his left eye turned out. There is no history of traumatism or syphilis. At the present time L.V. is 2-5; R.V. 2-3; both unimproved by glasses. There is complete left ophthalmoplegia externa, with the exception of the external rectus and possibly the superior oblique. Ptosis is most marked when patient is not fixing. Pupils react very sluggishly to light and accommodation. With the ophthalmoscope few fundus changes are visible, but the outlines of the papilla are not clean cut—it has a woolly appearance; fields of vision in both eyes are decidedly contracted for white and colors. Dr. Sanger Brown reports that, with the exception of the oculo-motor paresis and optic defects, there is nothing abnormal about his nervous apparatus. Dr. Elliot found saliva and urine practically normal and free from lead. ABRAHAMS.